JAMA Dermatology Clinicopathological Challenge

Annular Flaccid Pustules on the Trunk

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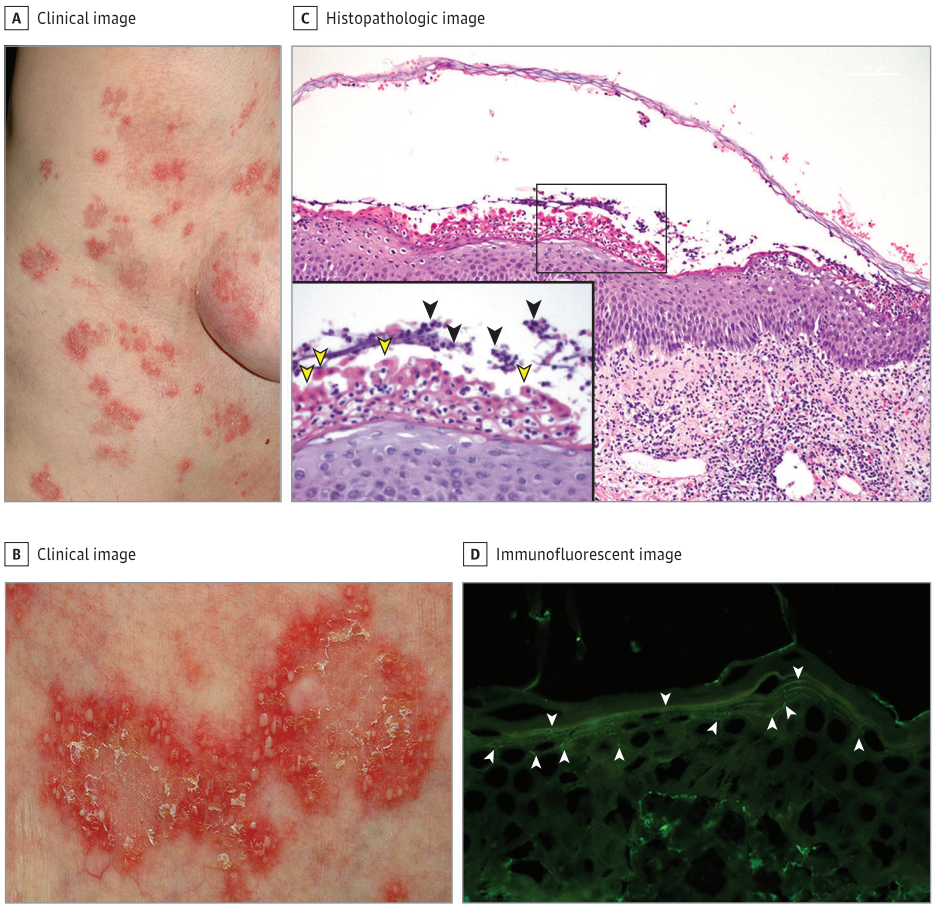

Figure. A and B, Clinical presentation showed flaccid pustules on scaly, erythematous plaques on the trunk. C, Subcorneal acantholysis of keratinocytes (yellow arrowheads) resulting in intraepidermal separation/blistering and an inflammatory infiltrate consisting of neutrophilic granulocytes (black arrowheads) (hematoxylin-eosin staining, original magnification × 100; inset original magnification × 400). D, Discrete subcorneal intercellular IgA depositions (white arrowheads) (direct immunofluorescence with FITC-labeled antihuman IgA antibodies, original magnification × 400).

A woman in her 50s presented for evaluation of itching skin lesions. The lesions had been present for months and increased gradually. Mucosae were unaffected. Treatment with topical steroids showed no lasting effect. Skin examination revealed multiple flaccid pustules on scaly, erythematous plaques on her trunk, with an annular and circinate pattern (**Figure**, A and B).

WHAT IS THE DIAGNOSIS?

A. Tinea corporis

B. IgA pemphigus

C. Duhring disease

D. Psoriasis pustulosa

Diagnosis

B. IgA pemphigus

Microscopic Findings

A biopsy revealed a subcorneal neutrophilic inflammatory infiltrate and acantholysis (Figure, C). Special stains and microbiologic examination findings were negative. Direct immunofluorescence microscopy revealed very subtle subcorneal intercellular IgA depositions (Figure, D). Immunoblots with recombinant desmosomal proteins (desmogleins 1 and 3 and desmocollins 1, 2, and 3) disclosed IgA-autoantibodies against desmocollin 3.

Discussion

IgA pemphigus is relatively rare and caused by IgA autoantibodies against desmosomal proteins.[1] It is traditionally subdivided into the subcorneal pustular dermatosis and the intraepidermal neutrophilic types.[2] Nevertheless, many cases of IgA pemphigus show atypical characteristics with IgA reactivity against various antigens in the epidermis and the alternative term IgA pemphigus spectrum has therefore recently been suggested.[3] The key clinical feature of IgA pemphigus is the presence of flaccid, sterile pustules on scaly, erythematous plaques. Because skin pustules are often caused by infections, the major differential diagnoses include impetigo (bacterial infection) and tinea (dermatophyte infection). If the microbiology is sterile, direct immunofluorescence is indispensable to distinguish IgA pemphigus from other sterile pustular dermatoses, such as psoriasis pustulosa, acute generalized exanthematous pustulosis, pemphigus foliaceus, and linear IgA dermatosis. The sample for direct immunofluorescence should be taken from perilesional skin and not from a lesion directly. The combination of subcorneal acantholysis in histologic analysis and intercellular IgA depositions in the upper epidermis in direct immunofluorescence—which can be subtle—is diagnostic for IgA pemphigus. Serological tests (indirect immunofluorescence or immunoblot) can be helpful to support the diagnosis, but frequently fail to identify the target antigen. Sensitivity of indirect immunofluorescence is reported to be only 50%, immunoblot techniques available in specialized centers can have a sensitivity of up to 60%.[2]

IgA pemphigus has been reported in association with malignant diseases[4-6] and other disorders, such as inflammatory bowel disease[7] and rheumatoid arthritis.[8] The disease is responsive to oral glucocorticoids, but often adjuvant therapy is required to allow for steroid tapering. Dapsone and colchicine are frequently used immunomodulators that inhibit recruitment of neutrophilic granulocytes to the skin. Further steroid-sparing agents are acitretin, and long-term immunosuppressants, such as azathioprine.[9] This patient did not improve on several immunosuppressive drugs and dapsone, but responded to treatment with colchicine.

ARTICLE INFORMATION

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Published Online: April 26, 2017. doi:10.1001/jamadermatol.2017.0901

Conflict of Interest Disclosures: None reported.

Additional Contributions: We thank R. Eming, MD, Philipps University Marburg, for help with the identification of IgA autoantibodies against desmocollin 3 and F. Schauer, MD, University of Freiburg, for clinical follow-up. We also thank the patient for providing permission to share her information. They were not compensated.

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